VOLUME-8, ISSUE-1, JANUARY-2019 • PRINT ISSN No 2277 - 8160

Original Research Paper

Anesthesiology

ELECTIVE CESAREAN SECTION IN SCIMITAR SYNDROME: ANAESTHESIA MANAGEMENT

Dr. A. Gangadhara Reddy M.D — Senior Consultant, Department Of Anaesthesia, Rural Development Trust And Hospital, Bathalapalli, Anantapur, Andhrapradesh,india.

Dr. S. Sonia Begum* — DNB Resident Department Of Anaesthesia, Rural Development Trust And Hospital, Bathalapalli, Anantapur, Andhrapradesh,india. *Corresponding Author

ABSTRACT

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies consisting of a partial anomalous pulmonary venous connection of the right lung (part or the entire right lung) to the inferior vena cava, right lung hypoplasia, dextroposition of the heart, and anomalous systemic arterial supply to the right lung. Anaesthesia management of pregnant patients with Scimitar syndrome is challenging due to coexisting congenital heart disease. The literature on anaesthesia management of patients with Scimitar syndrome undergoing Elective cesarean section is rare and hence authors wish to share their experience of anaesthesia management of a case of Scimitar syndrome posted for elective cesarean in a rural area hospital.

KEYWORDS : Scimitar Syndrome, Pregnancy

INTRODUCTION

The term scimitar syndrome or congenital pulmonary venolobar syndrome was first described by CATHERINE NEILL in 1960, describing a syndrome of partial anomalous pulmonary venous drainage of the right lung into the inferior vena cava, partial systemic arterial blood supply, and hypoplasia of the affected lung, with bronchial abnormalities and abnormal lobation. This is a rare anomaly with an incidence of approximately 1 to 3 per 100,000 live births.

The term scimitar syndrome was coined because of the radiographic appearance of the anomalous vein, which appears as a tubular opacity paralleling the right cardiac border resembling a curved Turkish sword or scimitar (scimitar sign). It is also called mirror image lung syndrome, hypo genetic lung syndrome, Halasz syndrome. Clinical presentation is of two types. Adult form and Infantile form with the infantile form presenting as severe pulmonary hypertension, cardiac failure, and a high mortality rate. Surgical intervention is required if there is a large left/right shunt exceeding 50%, resulting in pulmonary hypertension, heart failure or when there is lung sequestration and/or recurrent right-sided chest infections.

CARDIOPULMONARY CHANGES IN PREGNANCY

CARDIOVASCULAR CHANGES : Fall in total peripheral resistance by 6 weeks gestation to 40% by mid gestation. Circulatory underfilling activation of renin-angiotensin- aldosterone system .Total extracellular volume 16% by term. Plasma osmolality by 10mOsm/Kg as water is retained

The heart rate rises synchronously by 10-15 b.p.m. from 70 to 85 b.p.m. - stroke volume rises - cardiac output begins to rise by 35-40% in a first pregnancy and ~ 50% in later pregnancies

The blood pressure - slight drop in the 2nd trimester small fall in systolic, greater fall in diastolic B.P. opening of arterio-venous shunts at the placenta increased pulse pressure - supine hypotension syndrome in 8% of the women 2nd half of the pregnancy. Maternal hypotension occurs in the supine position due to pressure of the uterus on the inferior vena cava decreased venous return and cardiac output.

Noradrenaline - pressor response to angiotensin II reduced in normal pregnancy, unchanged to noradrenaline - plasma noradrenaline is not increased in normal pregnancy.

Pulmonary circulation - able to absorb high rate of flow without an increase in pressure - pressure in right ventricle, pulmonary arteries and capillaries does not change - pulmonary resistance falls in early pregnancy - progressive venodilatation + rises in venous distensibility + capacitance throughout a normal pregnancy.

RESPIRATORY CHANGES: Tidal volume rises by 30% in early pregnancy 40-50% by term Driven by. Fall in expiratory reserve and residual volume progesterone decrease the threshold increase the sensitivity of medulla oblongata to CO2.

Respiratory rate does not change the minute ventilation rises by a similar amount from 7.25L to 10.5L. Elevation of the diaphragm in late pregnancy causes dyspnea.

Carbon dioxide production rises sharply during the 3rd trimester as fetal metabolism increases. The fall in maternal P CO2 - allows more efficient placental transfer of CO2 from the fetus - results in a fall in plasma bicarbonate concentration (from 24-28 mmol/L to 18-22 mmol/L) fall in plasma osmolality venous pH rises slightly (from 7.35 to 7.38)

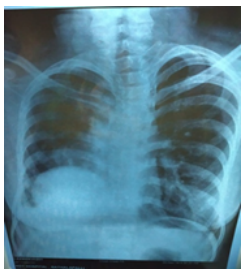
The increased alveolar ventilation small rise in PCO2 (from 96.7 to 101.8 mmHg). Rightward shift of the maternal oxyhaemoglobin dissociation curve (due to an increase in 2,3-DPG in erythrocytes) oxygen unloading to the fetus which has: - lower PCO2 (25-30 mmHg, 3.3-4 KPa) - marked leftward shift of the oxyhaemoglobin dissociation curve, (due to lower sensitivity of fetal haemoglobin to 2,3-DPG)

Increase of 16% in oxygen consumption by term. Fall in arterio-venous oxygen difference. Pregnancy places greater demands on the cardiovascular than the respiratory system

CASE REPORT

The present case is about a 20-year-old G2P1L1 with 32weeks of gestation with Scimitar syndrome who presented for safe institutional delivery. She was diagnosed as scimitar syndrome at the age of 10yrs. For her First baby was delivered by elective cesarean section not in this hospital 2yrs back under subarachnoid block, but details are not available.

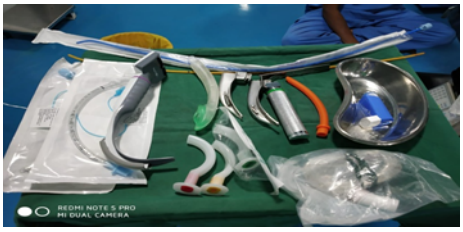
At the time of examination she gave past history of scimitar syndrome and on auscultation revealed reduced breath sounds over the right lung and heart sounds S1,S2 heard in right side of the chest. Echocardiography was showed Dextrocardia, Dilated RA/RV/PA, Mod TR, Mod PAH with Grade 2



Diastolic dysfunction with EF 65% Airway was normal.

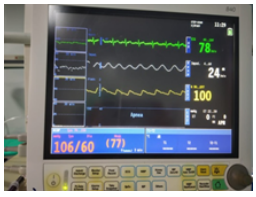
A multidisciplinary team consisting of an obstetrician, a visiting cardiologist and an anaesthesiologist closely monitored her during next 5weeks of her pregnancy. Cardiac function was assessed by echocardiogram before surgery .

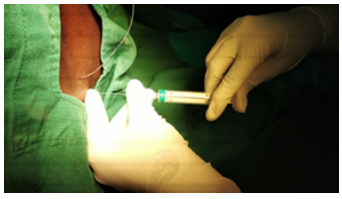
At 39-week gestation planned for cesarean section with past history of LSCS, not allowed spontaneous delivery. She presented to operation theater after taking of premedication with Inj.Ranitidine 50mg IV, Inj.Metachlopramide 10mg IV.



After all arrangements like emergency drugs tray, difficult airway cart including with video laryngoscopy we are planned Combined Spinal epidural anesthesia.

Preoperative vitals are monitored after securing IV catheter and placed 500ml 0.9% NS for preload. In sitting position patient was painted and draped under aseptic precautions.





With 18G Tuohy needle was used for epidural and 25G Quincke spinal needle was used for spinal anesthesia. For analgesia regime using a solution of 0.1% Ropivacaine and 100 mcg/ml of fentanyl was administered in 40 ml , providing 5 ml by continuous background infusion, after giving 0.5%H Bupivacaine 1.2ml with 25mcg Fentanyl in sub arachnoid space. Saline 0.9% was cautiously infused intravenously for maintenance of preload. After that Ringer Lactate solution was placed 60ml/hr.

Frequent clinical examination for signs of heart failure, with vital signs and SpO2 monitoring were performed during Combined spinal epidural and throughout cesarean section. She delivered Boy baby through incision after starting 5min of surgery.

The third stage was managed with 10 units intravascular oxytocin. No signs of cardiac failure were noted in the peripartum period. Her post-operative recovery was uneventful and for pain postoperative period continued epidural analgesia with same concentration of Ropivacaine with Fentanyl and she was discharged from hospital after 3days after epidural catheter was removed. Came back 7th day for suture removal without any complications.

DISCUSSION

Partial pulmonary venous return (PAPVR) is an uncommon anomaly accounting for 0.5-1% of all congenital heart diseases. The characteristic abnormality is PAPVR (partial or complete) of the right lung to the IVC, either at the junction of right atrium or below the level of diaphragm. Drainage into the right or left atria, superior vena cava, azygous, hepatic or portal vein can also occur. Atrial septal defects are the most common anomaly with PAPVR, but other congenital heart defects have also been described. Scimitar syndrome is a rare complex variant of PAPVR (3-5%), occurring in 1-3/100000 live births and consists of PAPVR, hypoplasia of the right lung and pulmonary artery, dextroposition of the heart, with right pulmonary arterial supply from the systemic circulation. Mostly, the pulmonary vein from the right lower lobe and occasionally the right middle lobe forms an abnormal 'Scimitar' vein that drains into the upper IVC below the level of the diaphragm. Consequently, a left to right shunt is established, with risk of right heart failure due to right ventricular volume overload.

Three forms of Scimitar syndrome are described. In the infantile form, patients become symptomatic early in life, developing severe PHT and cardiac failure, with cyanosis and failure to thrive at presentation. In the adult form, most patients are asymptomatic but the extent of symptoms depends upon the degree of shunting and the number of anomalous veins, associated valvular abnormalities and the presence of other concomitant cardiac or pulmonary disease. Recurrent pneumonia, mild dyspnoea on exertion and occasional haemoptysis are the most commonly reported symptoms. PHT is rare or mild and not associated with right heart failure when present. Scimitar syndrome in these patients is often discovered incidentally following a chest radiograph. The third form is associated with complex cardiac malformations like hypoplastic left ventricle or aortic arch obstruction, which are far more significant clinically. Some previously fit adults can present with infant form complications of PHT and right heart failure later in life. Previously asymptomatic pregnant patients may present with symptoms of volume overload due to the physiological increase of around 40% in blood volume by term; auto transfusion of 300-500 ml blood during each contraction in labour or iatrogenic fluid overload in the peripartum period.

Diagnosis is suspected on a chest radiograph showing a curved vascular shadow of the anomalous pulmonary vein descending towards the diaphragm along the right side of the heart (scimitar sign), along with lung hypoplasia and cardiac dextroposition. A transthoracic echocardiogram, MRI, contrast enhanced CT or cardiac catheterisation are more diagnostic.

Surgical correction in adults should only be considered in symptomatic patients or in asymptomatic patients with a pulmonary to systemic shunt blood flow ratio exceeding 1.5:1 due to their higher likelihood of developing PHT and right ventricular failure.

In our patient In the presence of significant PHT, anaesthetic management should focus on prevention of increased pulmonary vascular resistance (PVR) by avoidance of pain, hypoxemia, hypercarbia and acidosis. Fluid management may be difficult, as both hypovolemia and hypervolemia can prove detrimental. Crystalloids or colloids are both acceptable, but unmonitored fluid challenges may worsen right ventricular function and are not recommended.

Myocardial depression or marked reduction in systemic vascular resistance (SVR) and venous return should also be avoided and consequently epidural anaesthesia with incremental doses or a low-dose combined spinal-epidural anaesthesia may be preferable over spinal or general anaesthesia.

By reducing pain in labour, epidural anaesthesia prevents catecholamine release and a consequent increase in heart rate and SVR, and may be better tolerated than Entonox for labour analgesia. This is because nitrous oxide increases PVR and may worsen pre-existing PHT. Furthermore, use of vasoactive drugs like phenylephrine or ephedrine and uterotonic agents like oxytocin or ergometrine should be cautious in order to prevent acute haemodynamic effects.

Since the cardiac status in this patient was well-maintained throughout pregnancy, a cautiously extended epidural or a combined spinal-epidural was planned for any surgical intervention.

CONCLUSION

Anaesthetic management of patients with Scimitar syndrome for obstetric interventions should be guided by the mode of delivery planned; the degree of arterial-venous shunt present; the severity of

PHT and adequacy of ventricular function.

An epidural or a low dose combined spinal-epidural may be the technique of choice for obstetric surgical interventions or for labour analgesia in these patients.

It would be prudent to manage a parturient with deteriorating or complex pathology in a centre where facilities and expertise in dealing with adult congenital heart disease are available.

REFERENCES

1. Bell JD, Awonuga AO, Thompson ME, Ray CB, Devoe LD. Infantile scimitar syndrome complicating pregnancy. Int J Gynaecol Obstet. 2009;106:259–60. [PubMed]
2. Miyake M, Katayama S, Masaki K, Kubo H, Hirakawa S, Saji T. Scimitar syndrome and pregnancy: A case report. Congenit Anom. 2001;41:243.
3. Ranjith Baskar Karthekeyan, Yachendra, Siva Muthu Kumar, Suresh Rao, Mahesh Vakamudi, Balakrishnan Komarakshi, Richard Saldhana,Pneumonectomy in scimitar syndrome —is it correct? IJTCVS 2008;24:176- 179
4. Ajmer Singh, Neeraj Sharma Anaesthetic Management of Scimitar Syndrome: A Case Report"(http://www.theiaforum.org) July 2010
5. Moira A Hendrie, Deepak Mathur. Scimitar syndrome in pregnancy. Indian Journal of Anaesthesia 2014; 58:208-210
6. Kamler M, Kerkhoff G, Budde T, Jakob H. Scimitar syndrome in an adult: Diagnosis and surgical treatment. Interact Cardiovasc Thorac Surg. 2003;2:350–1. [PubMed]
7. Schramel FM, Westermann CJ, Knaepen PJ, van den Bosch JM. The scimitar syndrome: Clinical spectrum and surgical treatment. Eur Respir J. 1995;8:196–201. [PubMed]